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Letter to the Editor

CT-guided extracranial radiofrequency of multiple groups of cranial nerves for the treatment of compound Meige's syndrome

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TO THE EDITOR

As is well documented, compound Meige's syndrome is defined as segmental dystonia involving blepharospasm and involuntary twitching of oral and mandibular muscles [1]. As the disease progresses, it progressively spreads to the lower facial muscles, jaw, mouth floor, tongue, pharyngeal muscles, and respiratory muscles. At present, the most widely used treatment strategies are pharmacotherapy, botulinum toxin injections, and deep brain stimulation. Nevertheless, clinical understanding of compound Meige's syndrome is limited.

A 43-year-old man was admitted to the pain department of the authors' hospital for treatment with involuntary twitching on both sides of the face and neck for over 4 years. The patient was initially diagnosed with blepharospasm at another hospital and received injections of botulinum toxin near the orbital rim. While the patient experienced temporary relief, the duration of effectiveness steadily decreased from 4 months to approximately 1 month. Therefore, the patient underwent orbicularis oculi muscle resection at an external hospital, which alleviated the blepharospasm. However, masticatory and neck muscle spasms gradually worsened, leading to difficulty in opening the mouth and involuntary neck muscle contraction affecting head rotation.

A brain MRI (1.5 T, standard T1- and T2-weighted sequences, MR-angiography) displayed no evidence of vascular compression of the bilateral facial nerves. Consequently, "bilateral muscle spasm" was ruled out. Therefore, the patient was diagnosed with "compound Meige syndrome involving the V, VII, and XI cranial nerves."

This study was approved by the ethics committee of the affiliated hospital (approval number: LS2019-013), and the patient provided written informed consent. Briefly, the patient was placed supine on the CT table, and a coronal CT scan was performed. The puncture path was designed after locating the bilateral foramen ovale (FO). Next, two radiofrequency needles were punctured into the bilateral FO according to the designed path (**Fig. 1**) [2,3]. Following this, a motor nerve stimulation test was performed at a low frequency (2 Hz), and stimulation at 0.5–1 mA induced tremors in the bilateral mandibles matching the electrical stimulation frequency. The patient was sedated via intravenous injection of propofol 1 mg/kg and fentanyl 1 μg/kg, followed by 60 seconds of

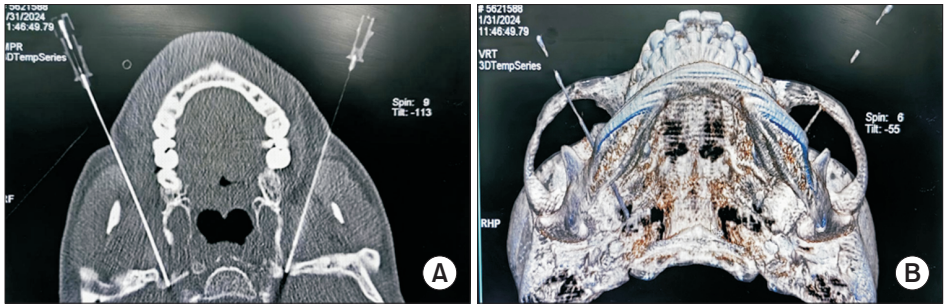

Fig. 1. Radiofrequency ablation of mandibular branch of trigeminal nerve by bilateral foramen ovale puncture under CT guidance to treat masticatory muscle spasm. (A) The successful image of bilateral foramen ovale puncture. (B) CT three-dimensional reconstruction image after successful puncture of the bilateral foramen ovale, with bilateral RF needle points located in the foramen ovale.

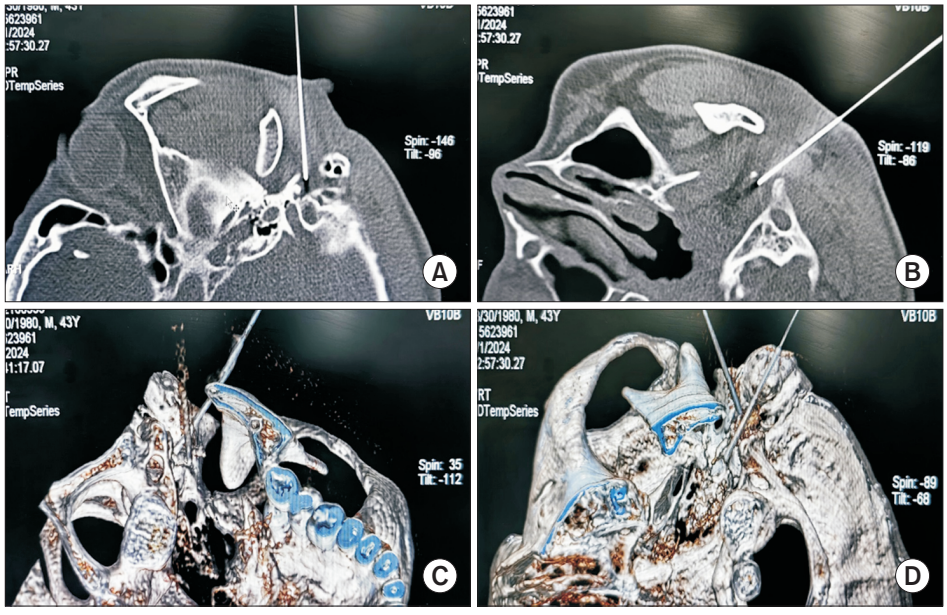

Fig. 2. (A) CT-guided radiofrequency ablation of facial nerve by right stylomastoid foramen puncture to treat eyelid spasms, lip muscle spasms, or platysma twitches. (B) CT-guided radiofrequency ablation procedure of the accessory nerve by the posterior edge of the styloid process at the level of the upper edge of the right atlas transverse process to treat involuntary head-turning movements. (C) CT three-dimensional reconstruction image after successful puncture of the right facial and accessory nerves with right RF needle points located in the stylomastoid foramen and the posterior edge of the styloid process. (D) CT three-dimensional reconstruction image after successful puncture of the left facial and accessory nerves with left RF needle points located in the stylomastoid foramen and the posterior edge of the styloid process.

continuous radiofrequency ablation at 70°C. Postoperative involuntary chewing was resolved, but this was accompanied by mild numbness and sensory reduction in skin areas innervated by the trigeminal nerve.

The next day, the patient was placed in the left lateral decubitus position on the CT table, and the right stylomastoid foramen was identified. Afterward, the puncture path was designed with the right stylomastoid foramen and the posterior edge of the styloid process at the level of the upper edge of the right atlas transverse process as targets [4,5], and two radiofrequency needles were inserted to the target according to the designed path (**Fig. 2A–C**). As anticipated, electrophysiological testing demonstrated that muscles innervated by the facial and accessory nerves displayed rhythmic contraction at the same frequency as the stimulation current. Thereafter, radiofrequency ablation was performed on the right facial nerve at 65°C for 8 seconds, resulting in symptoms of air leakage from the cheek and an inability to fully close the right eye. Subsequently, radiofrequency ablation was

conducted on the right accessory nerve at 75°C for 60 seconds [6–8]. Subsequently, the left side was treated in the same way (**Fig. 2D**).

On the one hand, eyelid spasms, mouth twitching, and head and neck torsion were resolved, with no residual mouth asymmetry. On the other hand, slight air leakage was detected during cheek puffing, accompanied by no involuntary movements of the tongue, and no difficulty in swallowing or phonation.

Six months after discharge from the hospital, video and telephone follow-ups unveiled no recurrence of involuntary facial or neck twitching. In addition, the symptoms of facial paralysis had been resolved, and numbness in the mandibular branch of the trigeminal nerve and chewing ability had improved. More importantly, the patient did not require medications and reported significant improvement in quality of life.

As a segmental craniocervical dystonia, the mechanism underlying MS remains to be elucidated. The analysis of current treatment techniques for Meige syndrome (MS) reveals four main approaches: severing or destroying the effector muscles, blocking signal transmission at the neuromuscular junction, interrupting motor signal transmission along cranial nerves, or interfering with or modulating the generation of motor signals in the central motor nuclei. Local botulinum toxin injections work by blocking the transmission of abnormal motor signals at the neuromuscular junction, acting directly on the effector. Deep brain stimulation (DBS) treats MS by blocking or interfering with the generation of abnormal motor signals in the subthalamic nuclei, addressing the "pacemaker" or signal source. Based on the therapeutic principle of inhibiting motor signal transduction, extracranial partial radiofrequency ablation of the responsible nerve was performed, which not only effectively blocked the conduction of abnormal motor signals along the nerve but also preserved the conduction of some normal voluntary motor signals. Overall, this approach attenuates the symptoms of MS and concurrently retains partial physiological functions of cranial nerves.

As the authors performed partial radiofrequency ablation on the responsible cranial nerves, there remains a possibility of recurrence. Post-procedural complications may include mild facial paralysis following facial nerve ablation and square shoulder deformities following accessory nerve ablation. Although there was no difficulty in mouth opening after bilateral radiofrequency ablation of the mandibular branch of the trigeminal nerve, residual nerve sensory loss and chewing fatigue were observed, which persisted for a prolonged period. Taken together, this approach is anticipated to emerge as a safe and effective method to address the clinical challenge of MS treatment [9,10].

DATA AVAILABILITY

Data sharing is not applicable to this article as no datasets were generated or analyzed for this paper.

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The authors would like to dedicate this article to the patients who have suffered from Meige's syndrome.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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